

*Prikaz slučaja /
Case report*

TRANSFORMATION OF
DERMATOFIBROSARCOMA
PROTUBERANS IN VERY AGGRESSIVE,
EXTREMELY FAST GROWING AND
DESTRUCTIBLE TUMOR: *A Case Report*

TRANSFORMACIJA
DERMATOFIBROSARKOMA
PROTUBERANSA U VEOMA AGRESIVAN,
IZUZETNO BRZO RASTUĆI I
DESTRUKTIVAN TUMOR - *Prikaz slučaja*

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Ključne reči

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dermatofibrom,
margine resekcije tumora

Key words

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Abstract

In this article presented a 42-year-old patient who underwent multiple operations for locally aggressive dermatofibrosarcoma protuberance of the face at the Maxillofacial Surgery Clinic of the Military Medical Academy in Belgrade. A patient with an extremely aggressive form of dermatofibrosarcoma protuberance of the middle third of the face who was operated on seven times in two years is presented. He was operated four times for an extremely large tumor (size about 200x120cm), which first grew to that size within just one month and recurred within a short period of time after each operation. Dermatofibrosarcoma protuberans is a locally aggressive tumor and can have a rapid and destructive course, which generally has no metastatic potential, and surgical treatment is considered the best modality of treatment. Due to the pronounced local aggressive behavior, this form of tumor can often be completely removed surgically. That is why a wide local excision with margins of 3-5 cm towards healthy tissue is suggested today, as well as resection of deep structures, including bone. In patients with recurrent tumors, the best result that can contribute to successful surgical treatment involves wide resection margins of soft and bone tissues, regardless of functional and aesthetic consequences.

INTRODUCTION

Dermatofibroma is universally held to be a benign tumor. Dermatofibrosarcoma protuberans (DFSP) belongs to the group of fibrohistiocytic tumors. It's indolent, low-grade sarcoma that grows in an infiltrative fashion, presented as asymptomatic cutaneous nodule in middle adult life³ with a marked tendency for local recurrence, but it rarely metastasizes^{1,2}. Dermatofibrosarcoma protuberans accounts

for less than 5% of adult soft tissue sarcomas and less than 1% of all malignant tumors of the head and neck⁴. It dominantly arises on the skin of the trunk (50-60%), proximal extremities (20-30%) and the head and neck (10-15%)^{5,6}. So, 90% or more of DFSP a characterized with translocation between chromosomes 17 et 20⁶⁻⁸. Because of its seemingly benign appearance, misleading surgeons and patients, treatment frequently is delayed and/or inadequate and in the 50%

of cases it may exceed 10 years from the first signs of illness till first resection^{8,9}. DFSP represents about 5% of head and neck sarcomas and because of that it's usually underscored⁵⁻⁹. Treatment for DFSP is surgical resection. Standard wide excision with a width of 1 cm around the primary tumor would have left microscopic residual tumor in 70.7%; a width of 2 cm, 39.7%; 3 cm, 15.5%; and 5 cm, 5.2%, even more, in some cases tumors were not completely removed with 10 cm margins^{1,8-11}. Microscopic exam shows multiple finger-like projections of neoplastic cells on the tumor's periphery that can spread laterally or deeply into underlying fascia and muscle even 3 cm from the main tumor¹²⁻¹⁴. This is the reason why most surgeons recommend wide excision with more than 3 cm grossly uninvolved margin of skin and underlying deep fascia¹⁵. This work presents a case of dermatofibrosarcoma protuberans of the cheek, three years after initial excision of dermatofibroma.

CASE REPORT

A 42-years-old man noticed discreet, painless left cheek soft tissue lesion in December 2001, about 1 cm in diameter. Lesion was smooth, elastic and painless, well differentiated from the surrounding tissue with no alteration in skin colour. Lesion has been fixated for the skin and movable in regard to the underlying tissue. He underwent surgical excision and defect was primary closed. First pathohistological result

indicated that lesion was dermatofibroma. Three years later, in June 2004, he was operated because of the recurrence at the site of excision and pathohistological result indicated dermatofibrosarcoma protuberans. In the same month, in the Clinic for Maxillofacial Surgery of Military Medical Academy in Belgrade, patient underwent excision of surrounding tissue, partial resection of left maxillary bone and nasal bones on the same side and the defect have been reconstructed with local flaps. Pathohistological examination was the same as the last one. In the November 2005 wide surgical excision was performed on the left and right cheek and superior lip. Pathohistological examination was the same as the last one. In March 2006 he was operated because of residual tumor on the left side of the face. In the September 2006 (Figure 1), huge residual tumor, spreading from the midface on the right side and penetrating right orbit, was the reason for the exenteration of the right orbit and complete resection of right maxillary bone and partial resection of left maxillary bone without reconstruction (Figure 2). Percutaneous endoscopic gastrostomy has been used because the patient was unable to feed himself normally. Pathohistological examination was the same. Because of rapidly growing tumor, that achieved size of the patient head during three months (Figure 3), tracheotomy and palliative surgery was performed in January 2007 with no reconstruction and the defect involved midface, right orbit and perimandibular



Figure 1. Before operation in September 2006.

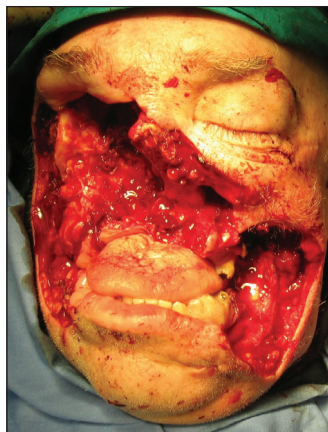


Figure 2. After operation in September 2006.



Figure 3. Before operation in January 2007.

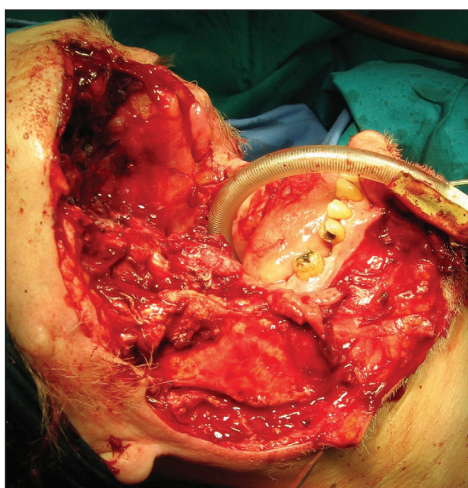


Figure 4. After operation in January 2007.

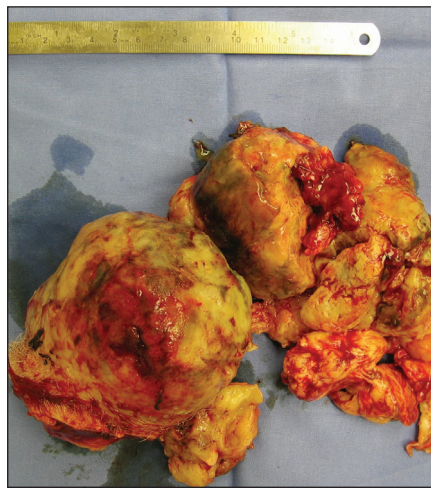


Figure 5. Surgical specimen after operation



Figure 6. Before operation in February 2007. in January 2007.

region on the both sides (Figure 4 and 5). Tumor had involved all structures of the facial bones, left eye, not penetrating into the cranium, without local or distant metastasis. Only one month after that patient underwent another paliative surgery with the tumor involving frontal region, midface, perimandibular region, leaving only the tongue intact (Figure 6). It was even more bigger than the last time.

DISCUSSION

DFSP represents less than 5% of all sarcomas and less than 1% of all malignant tumors of the head and neck.¹⁻³ 10-15% of all DFSP have been reported in head and neck region.⁶ We presume that even at the time of the first excision the pathohistological result was DFSP and not dermatofibroma because it is well known that is difficult to distinguish these two entities but the main difference is in presence of irregular, interwoven fibrocellular fascicles composed of uniform spindle-shaped cells and variable amounts of collagen, which produced a storiform appearance.^{4,12}

DFSP is unusual, slow growing, painless sarcoma with low grade malignancy arising in dermis and subcutaneous tissue presented as cutaneous nodule. It is locally very aggressive but it rarely metastasizes.^{1,5} Also, rapidly growth and aggressive behavior of this tumor in a last few month could be associated with amplification of fibrosarcomatous portion. The most significant factor that predicts outcome for DFSP is extent of resection. Wide resection is not always tenable, particularly on the face, where an extensive resection may entail significant functional and/or cosmetic deficit. That is the reason because of which local recurrence rate is higher in head and neck region. Adequate initial resection is important because multiple local recurrences predispose to distant disease spread. Overall 5-year survival rates after

adequate surgical excision are 93% to 100%. Minimum gross resection margin that ensure local control is undefined, but many authorities would agree that a margin of normal tissue 3-5 cm from gross tumor boundary including skin, subcutaneous fat and underlying fascia must be achieved to ensure adequate excision.^{5,12-14} To optimize local control of DFSP, some physician applied Mohs micrographic surgery to treat DFSP but this technique adequate for large and recurrent tumors that are very aggressive¹⁵.

CONCLUSION

DFSP is rare tumor in head and neck region, it is asymptomatic and it rarely metastasizes. Overall survival after adequate surgical excision is excellent. Deep lesions requires precise microscopic examine of resection margins. In our opinion it is necessary to see amount of fibrosarcomatous portion in tumor. Minimum resection margin must be 3-5 cm or more. Resection of deep structures in recurrent tumors that assure successful surgical treatment must include and bone structures regardless of cosmetic and functional defects. Often controls for many years are necessary.

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Sažetak

U radu je prikazan 42- godišnji pacijent koji je u više navrata operisao lokalno agresivni dermatofibrosarkoma protuberans lica u Klinici za Maksilofacijalnu hirurgiju Vojnomedicinske akademija u Beogradu. Prikazan je pacijent sa izuzetno agresivnim oblikom dermatofibrosarkoma protuberans srednje trećine lica koji je operisan sedam puta a dve godine. Četiri puta je operisan zbog izuzetno velikog tumora (veličine oko 200x120cm), koji je prvi put nastao do te veličine u roku od samo mesec dana i isti se ponovo javio u kratkom vremenskom periodu nakon svake operacije. Dermatofibrosarkom protuberans je lokalno agresivan tumor i može imati brz i destruktivan tok, koji uglavnom nema metastatski potencijal te se hirurško lečenje smatra najboljim modalitetom lečenja. Zbog izraženog lokalno agresivnog ponašanja često se ovaj oblik tumora može hirurški u celosti ukloniti. Zato se danas predlaže široka lokalna ekscizija sa marginama od 3-5 cm prema zdravom tkivu, kao i resekcija dubokih struktura, uključujući i kost. Kod pacijenata sa rekurentnim tumorom najbolji rezultat koji može doprineti uspešnom hirurškom lečenju podrazumeva široke margine resekcije mekih i koštanih tkiva bez obzira na funkcionalne i estetske posledice

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